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Case Report

Free Fibula Flap Mandible Reconstruction for Oral Obstruction Secondary to Giant Fibrous Dysplasia

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Fibrous dysplasia is a benign, proliferative disease of bone. The fibrous, bony lesions were first described in 1891 by von Recklinghausen,[1] who included two such cases in a report on the osteitis fibrosa cystica of hyperparathyroidism. An association with cutaneous and endocrine pathology was not noted until two reports of six cases in 1937. Five patients were described[2] with unilateral, fibrous, bony lesions, brown skin pigmentation in a similar distribution, precocious puberty in females, and early union of the epiphyses. A further case with a similar constellation of findings and hyperthyroidism also was reported.[3] Based on these two latter reports, that association has been termed the *McCune-Albright syndrome.* In 1938, Lichtenstein[4] characterized the distinct condition of multiple osseous lesions as "polyostotic fibrous dysplasia." He reviewed the clinical, radiographic, biochemical, and pathologic features of this entity. A detailed review of monostotic fibrous dysplasia appeared in 1946.[5]

An early, detailed study[6] described the pathology as consisting of replacement of cancellous bone by proliferative fibrous tissue admixed with disorganized trabeculae of woven bone. Expansion and distortion occur within the thinned subperiosteal cortex. The gross appearance is gray-white or gray-red, with a gritty texture. Histologically, mature connective tissue is dominant; in it, the irregular trabeculae of woven bone are scattered, and occasional foci of calcification or cartilaginous tissue may be found.[7] While fibrous dysplasia is believed to be a benign disorder, rare sarcomatous degeneration also has been implicated. Reports in the literature[8–10] have described 30 such cases, with an incidence of 0.4 percent. Radiation therapy appears to be an important risk factor, since 13 of these 30 patients had received such treatment.

The etiology of fibrous dysplasia remains unknown. Various authors[2,4] have attributed the disease to embryologic, neurologic, or congenital origins; others have argued against a congenital origin, favoring a "disturbance of the normal reparative processes" following trauma.[5] Edgerton and colleagues[11] proposed that fibrous dysplasia represents a congenital hamartoma.

Fibrous dysplasia appears to occur equally in males and females.[12–15] Lesions typically begin during childhood, are slow growing, often asymptomatic, and may decrease or cease growth after adolescence. The belief that fibrous dysplasia becomes quiescent after puberty has been widely accepted,[16] but examples of continued or accelerated growth later in life are scattered throughout the literature. The most common sites of involvement are the ribs, femur, tibia, and craniofacial skeleton. Within the craniofacial region, the maxilla is most often affected, followed by the mandible and calvarium. Disease is monostotic in 70 percent of patients, polyostotic in 20 percent of patients, and polyostotic with endocrinopathies and cutaneous pigmentation in 3 to 5 percent of patients. Craniofacial involvement occurs in 10 percent of monostotic disease and in 50 to 100 percent of polyostotic disease.

Extensive data regarding the signs and symptoms of craniofacial involvement are found in

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several reviews[11,13–15,17,18] of large series of patients. These include painless swelling with visible deformity (most common), headache, anosmia, orbital dystopia, diplopia, proptosis, decreased visual acuity, epiphora, strabismus, facial paralysis, hearing loss, tinnitus, sinusitis, nasal obstruction, epistaxis, malocclusion, and interference with mastication and speech. Radiographs reveal a ground-glass appearance of the bony lesions, classified into subtypes by Fries.[17]

Case Report

A 34-year-old woman presented with oral obstruction secondary to giant fibrous dysplasia of the mandible, a component of polyostotic fibrous dysplasia with severe craniofacial involvement. Growth of the left side of the mandible had progressed to the point where she had extreme difficulty in eating and swallowing.

The patient was the product of a full-term, uncomplicated pregnancy of a nonconsanguineous marriage. At birth, she had pigmentation on her back, right leg, and left clavicular area. Her mother first noted enlargement of the maxillary alveolus and left facial hypertrophy at the age of 5 years. At 7 years of age, a diagnosis was made of polyostotic fibrous dysplasia (McCune-Albright syndrome). The patient underwent menarche at 10 years of age. There was no family history of fibrous dysplasia or von Recklinghausen's disease.

Thirteen partial resections of the facial skeleton, including the mandible, were performed during childhood and adolescence; nevertheless, the disease progressively worsened. The patient was referred to the Center for Craniofacial Disorders at Montefiore Medical Center when she was 23 years old. At that time, her physical examination was significant for macrocranium, hypertelorism, and dysplasia of the maxillae and mandible (Fig. 1). Severe open-bite malocclusion was present, with missing maxillary teeth, including all the central and lateral incisors. All canthal and cranial measurements were above the 98th percentile, and endocrine evaluation was within normal limits. Over a 4-year period, seven partial resections were performed at Montefiore for disease of the orbits, zygoma, maxillae, and mandible.

Subsequent to these procedures, the patient sought consultation at another center, where radical, multistage surgery was proposed, only to be canceled after the patient was diagnosed with hyperthyroidism. She returned home, where she was treated with radioactive iodine; in addition, a large ovarian cyst was resected. She then returned to Montefiore with severe enlargement of the left mandible. Extraoral resection of a 15 × 13 × 4 cm mass of fibrous dysplasia was performed, preserving the inner cortex of the mandible. Rapid regrowth occurred over the next 2 years, culminating in oral obstruction. At this time, physical examination revealed a highly dysmorphic appearance with panfacial involvement, including gross enlargement of the calvarium, hypertelorism, maxillary hyperplasia, and a 15 × 12 cm mass extending from the mandibular symphysis to the left ramus (Fig. 2). Laboratory values were significant for elevation of serum alkaline phosphatase to 562 units/liter (normal adult 30 to 100 units/liter).

Skeletal radiographs revealed extensive polyostotic fibrous dysplasia of the calvarium, facial bones, left clavicle, multiple ribs, and left fibula (Fig. 3). CT scan was remarkable for involvement of the calvarium, skull base, clivus, cribriform plate, orbits, maxillae, and mandible (Fig. 4). There was stenosis of the optic canals, with atrophy of the optic nerve bilaterally; there was also stenosis of the external auditory canals and of the nasal passages. Panorex views were unobtainable because of the enormous size of the mandibular mass.

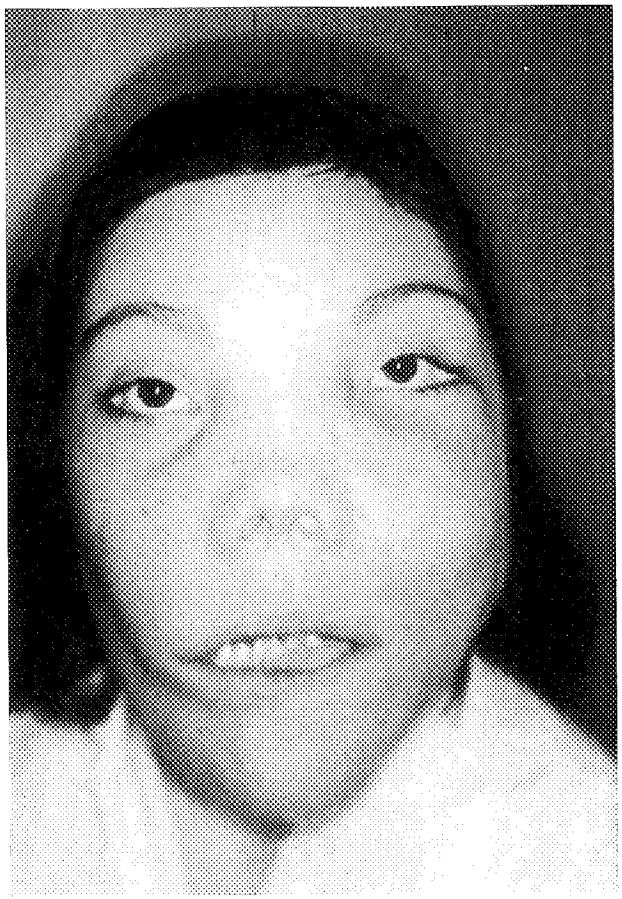

Fig. 1. Dysmorphic appearance at age 23, on presentation to Montefiore Medical Center.

At operation, a tracheostomy was required for general anesthesia. An extraoral mandibulectomy was performed from the right parasymphysis to the left ramus. The facial artery and vein had been ligated during previous surgery. The greater saphenous vein was harvested, and a preliminary arteriovenous fistula was established from the left external carotid artery to the left internal jugular vein, for these vessels to serve as recipients for a free bone flap. The right fibula, not radiographically involved in the fibrous dysplasia, was then isolated on its peroneal blood supply. Fibular osteotomies were performed in situ, based on a template of the mandibular defect, and secured with rigid miniplate fixation. The fibula was then transferred to the defect and secured to the native mandible with rigid miniplate fixation, and microvascular anastomoses were performed to the vein grafts provided by division of the arteriovenous fistula. The resected specimen measured 12 × 10 × 8 cm and weighed 520 gm. Histopathologic studies confirmed fibrous dysplasia.

The fibula flap healed, thus restoring mandibular continuity (Fig. 5) and relieving the oral obstruction. Oral intake was now possible, with unobstructed access to the oropharynx (Fig. 6). Preoperatively, no oral intake was possible, and the patient had refused placement of a gastrostomy tube. While

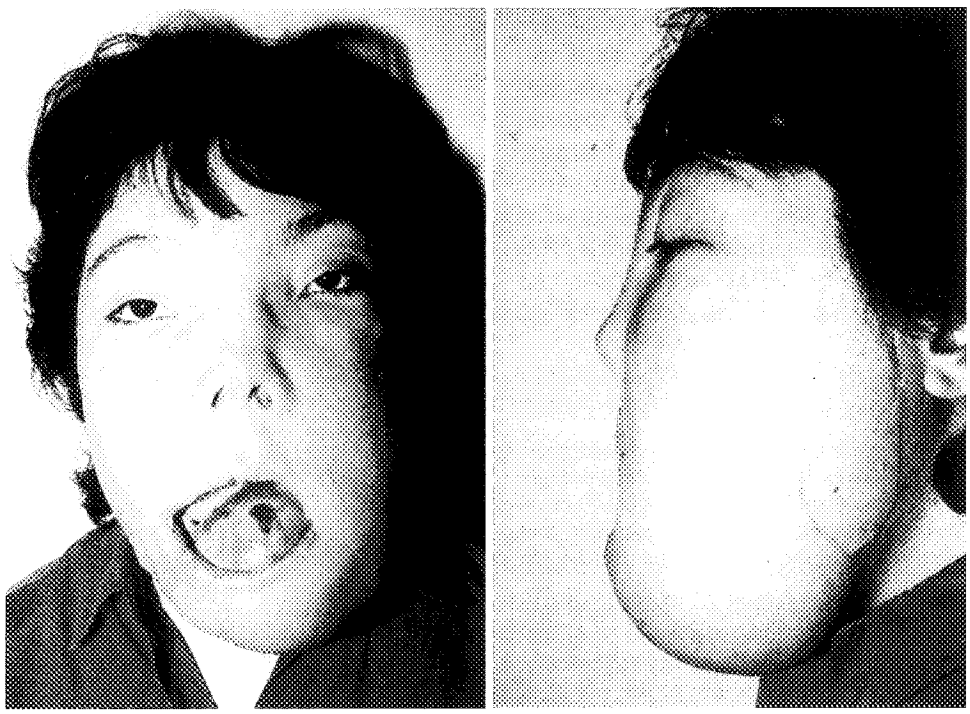

FIG. 2. Anteroposterior and lateral views at age 33; oral obstruction necessitated urgent treatment.

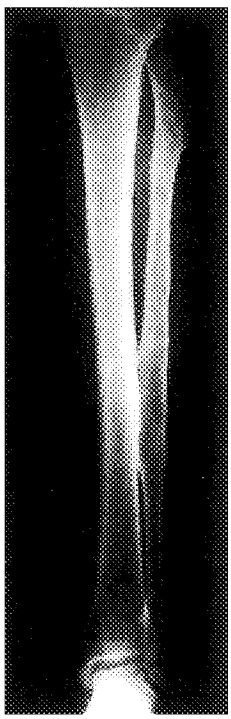

FIG. 3. Left fibula involved with fibrous dysplasia.

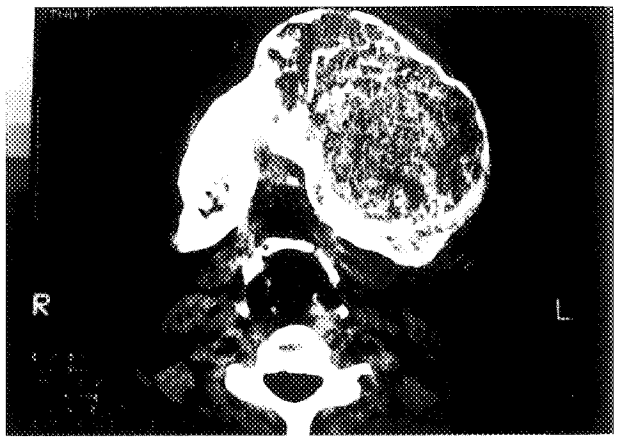

FIG. 4. Preoperative axial CT scan at level of mandible.

the patient remains highly dysmorphic (Fig. 7), it was never intended to improve aesthetics or reconstruct dentition and occlusion at this time. Now, with the patient 35 years of age, the fibrous dysplasia continues to grow throughout the craniofacial skeleton, except for that portion of the mandible reconstructed with the fibula. She has since undergone further surgery to reduce right maxillary hypertrophy, which impacted on the mandible and produced early closure, as well as obstructed the right nasal passage. In addition, a later surgery, unrelated to the fibula, allowed direct visualization of the fibulomandibular junction (Fig. 8). This provided the opportunity to show clearly that the fibula flap had healed to dysplastic native mandible.

DISCUSSION

Indications for surgical treatment of craniofacial fibrous dysplasia include cosmetic deformity, pain, pathologic fracture, to rule out malignancy, and functional considerations of vision and hearing, sinus and nasal obstruction, epistaxis, malocclusion, and mastication. The traditional, conservative management (in hope that the disease activity would burn out) has

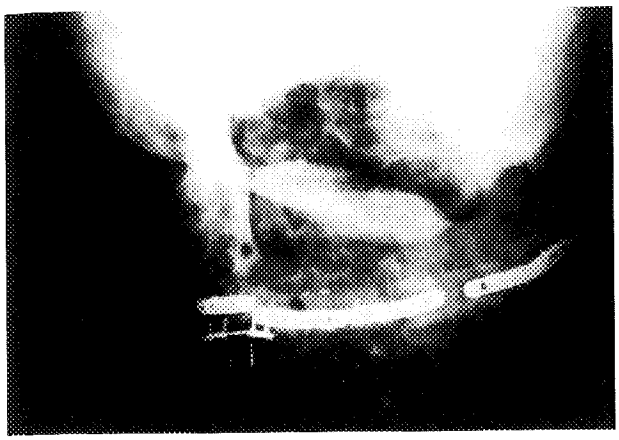

FIG. 5. Postoperative x-ray with fibula flap healed to adjacent dysplastic mandible.

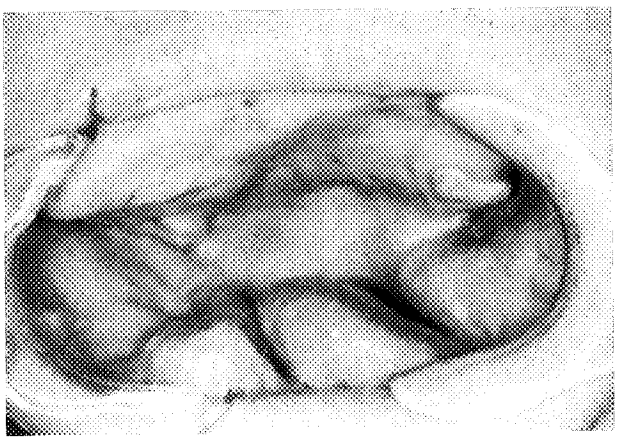

FIG. 6. Postoperative unobstructed access to oral cavity.

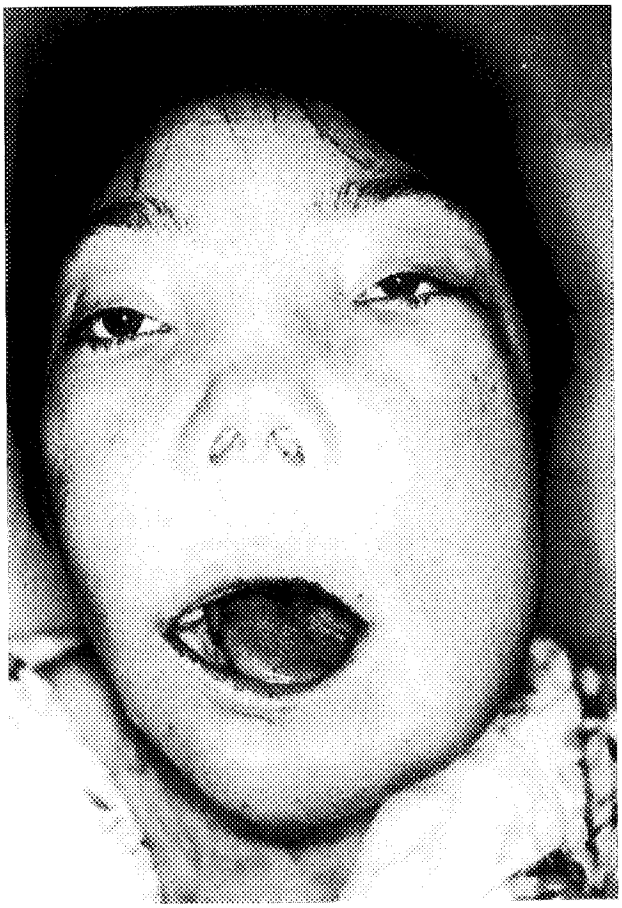

FIG. 7. Postoperative appearance.

evolved into a variety of aggressive treatment strategies. The armamentarium of shaving, curettage, and partial and total resections with bone-graft reconstruction has been expanded to include new techniques.

An earlier report[19] on 5 patients with frontoorbital fibrous dysplasia cited radiographic[14] and pathologic[20] studies confirming the continued progression of the disease beyond puberty and advocated radical resection with rib grafting. A subsequent report[11] on 23 patients cited a 25 percent incidence of local recurrence following conservative partial resection and recommended early surgery for cosmetic or functional correction. The authors of the latter report highlighted a technique to "remove, remodel, and replace" dysplastic bone with autogenous bone grafts, demonstrating the clinical cessation of dysplastic activity through the use of grafts. In addition, methyl methacrylate implants were used for large defects. Both these techniques avoided the donor-site morbidity of relatively large bone-graft harvests. More recently, Chen and Noordhoff,[18] in a series of 28 patients, divided the craniofacial skeleton into four zones based on functional, technical, and cosmetic considerations and stratified treatment accordingly. Total excision with primary bone grafting was performed in the fronto-orbital, zygomatic, and upper maxillary regions. Conservative excisions were performed for hair-bearing skull, central cranial base, and tooth-bearing bones. Optic nerve decompression was used for symptomatic nerve compression.

Although reports on giant fibrous dysplasia of the mandible are sparse in the literature, an evolution in the sophistication of treatment has occurred. While osteotomies for repositioning[18] and decompression of the mandibular canal[21] are useful, the mainstay of treatment has been hemimandibulectomy or subtotal mandibulectomy. Our patient represents an extreme case of pathologic growth, demonstrating the failure of conservative resection and indicating the need for aggressive resection.

Nonmicrosurgical reconstruction for these large mandibular defects historically employed bone grafting or prosthetic replacement. In

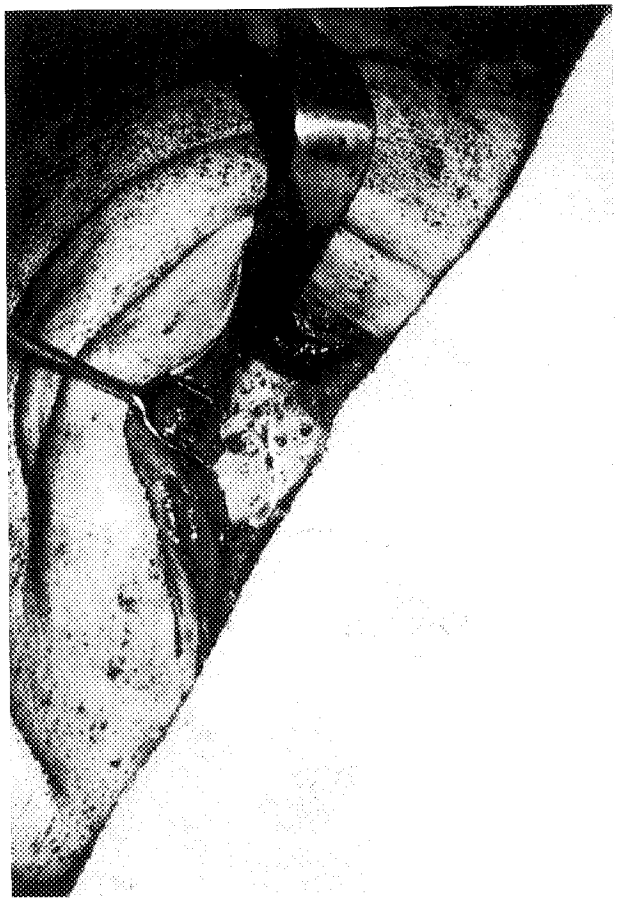

FIG. 8. Healed fibulomandibular junction at 18 months.

1955, Lewin[22] reported on four cases of mandibular fibrous dysplasia, two of which were gigantic. One of these patients had a life-threatening oral obstruction, requiring tracheostomy prior to resection. An immediate metallic rod reconstruction ultimately extruded and was replaced by delayed bone grafts. Ten years later, Manchester[23] described a case of giant fibrous dysplasia, with reconstruction using an iliac bone graft. A subsequent report concerned two cases of giant fibrous dysplasia of the mandible,[24] one treated with an iliac bone graft and one with a prosthetic tray filled with particulate graft. A later paper[25] demonstrated the use of a methacrylate resin hemimandible implant.

Microsurgical bone-flap reconstruction for fibrous dysplasia of the mandible has rarely been reported. A series of 12 mandibular reconstructions with free fibula flaps included one patient with "ossifying fibroma," in whom resection for persistent pain was carried out.[26] Chen and Noordhoff[18] included in their series one patient with a painful recurrence of fibrous dysplasia, with reconstruction using a free iliac crest flap.

Unlike these examples, our patient underwent resection for gigantic disease with oral obstruction and immediate reconstruction with a free fibula flap. Microsurgical free transfer of the fibula was first introduced by Taylor et al.[27] in 1975. Subsequently popularized for mandible reconstruction,[26,28] the advantages of this donor site include ample length of bone, the ability to support multiple osteotomies, easy contouring, a two-team approach, and low donor-site morbidity. In our application, the fibula was employed to restore bony continuity of the mandible in a setting of subtotal palliative resection. With rigid fixation, the fibula healed to dysplastic bone. To date, there has been no dysplastic growth of the fibula itself; however, dysplastic mandible adjacent to the fibula continues to grow. This case demonstrates not only the virulent regrowth capacity of fibrous dysplasia but also the usefulness of microsurgical bone-flap reconstruction in giant fibrous dysplasia of the mandible.

SUMMARY

Fibrous dysplasia is a disorder of bone that may be associated with endocrinopathies and skin pigmentation. The pathologic, proliferative expansion and distortion of the skeleton is of unknown etiology. Craniofacial involvement that includes the mandible can exhibit gigantic disproportions and dysfunction. Treatment has evolved to include more aggressive strategies of resection and sophisticated reconstructive techniques. The reported case is noteworthy for the unrelenting growth of craniofacial fibrous dysplasia in an adult female with endocrinopathies, progressing to oral obstruction that required urgent treatment utilizing immediate free bone-flap reconstruction. The free fibula flap was employed to restore mandibular continuity after palliative subtotal mandibulectomy. Bony healing to dysplastic tissue occurred in the remaining mandibular segment. This case illustrates that fibrous dysplasia has the capacity for virulent regrowth subsequent to conservative resection. Defects following radical surgery for giant fibrous dysplasia of the mandible can be reconstructed with a microsurgical bone-flap technique.

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